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Clinical Notes

Orthognathic Surgery in Craniomaxillofacial Fibrous Dysplasia

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In craniomaxillofacial fibrous dysplasia, jaw involvement often causes facial asymmetry, an occlusal cant, and loss of teeth. Although conservative management of fibrous dysplasia affecting the jaws is widely practiced, orthognathic surgery is indicated in such cases to restore occlusion and correct dentofacial deformity brought on by the disease process. Since 1981, the Craniofacial Center at Chang Gung Memorial Hospital in Taiwan has treated a total of 84 patients with craniomaxillofacial fibrous dysplasia. Of these, 55 (65%) had fibrous dysplasia affecting the jaws (Zone 4). Between 1988 and 1997, orthognathic surgery was performed on 1 male and 4 female patients with fibrous dysplasia involving the teeth-bearing jaws. One patient had localized fibrous dysplasia that involved the mandible. The other 4 patients had polyostotic craniofacial involvement of Zones 1, 2, or 3 and 4A. The patient with isolated mandibular involvement and 2 patients with maxillary fibrous dysplasia had single-jaw surgery. The other 2 patients with maxillary involvement required simultaneous two-jaw surgery to correct the dentofacial deformities resulting from the disease process. Follow-up ranged from 12 months to 9 years. All the patients had stable occlusion, good facial aesthetics, and no further recurrence after surgery. The long-term stability of the achieved occlusion and facial appearance confirms that adequate healing in fibrodysplastic bone is to be expected using the standard fixation.

Key Words: Orthognathic surgery, fibrous dysplasia

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Fibrous dysplasia is a congenital, metabolic, nonfamilial, non-neoplastic disease of the bone. It may be monostotic (involving one bone) or polyostotic (involving more than one bone) in nature, and at times is associated with skin pigmentation or endocrine abnormalities, such as McCune-Albright syndrome first described in 1937.[1] The term *fibrous dysplasia* was coined by Lichtenstein[2] in 1938, but Thoma[3] is credited with first using the term *facial fibrous dysplasia* in 1954.

In craniomaxillofacial fibrous dysplasia, craniofacial involvement occurs in 27% of monostotic and up to 50% of polyostotic patients.[4] Frontal, orbital, and sphenoidal bones are frequently involved, causing visual disturbances, proptosis, orbital dystopia, and often grotesque facial asymmetry. Jaw involvement may involve a solitary lesion, but nonetheless is more common in the maxilla than in the mandible.[4]

Indications for surgery are determined by the site, rate of growth, and involvement of contiguous structures. Advances in craniofacial surgery have allowed us to move beyond conservative excision and recontouring to more aggressive radical surgery combined with primary reconstruction using bone grafts. We have previously redefined the treatment principles of craniomaxillofacial fibrous dysplasia based on our classification of the craniomaxillofacial skeleton into four zones.[4]

Fibrous dysplasia involving the jaws (Zone 4) often gives rise to malocclusion, facial asymmetry, and jaw disproportion. Although conservative management of fibrous dysplasia affecting the jaws is widely practiced, orthognathic surgery is indicated in such cases to restore occlusion and correct dentofacial deformity brought on by the disease process. At the Craniofacial Center of Chang Gung Memorial Hospital, we have performed orthognathic surgery where indicated on five such patients. These five cases demonstrate not only the feasibility of orthognathic surgery in the management of dentofacial de-

formities caused by fibrous dysplasia, but also the uncomplicated healing of the osteotomies performed.

Patients and Methods

Since 1981, the Craniofacial Center at Chang Gung Memorial Hospital in Taiwan has treated a total of 84 patients with craniomaxillofacial fibrous dysplasia. Of these, 55 patients (65%) have fibrous dysplasia affecting the jaws (Zone 4). Among them, 44 patients (80%) had involvement of the maxilla, 8 (14.5%) had fibrous dysplasia of the mandible, and 3 (5.5%) had disease affecting both jaws. Of these 55 patients, 22 (40%) had isolated fibrous dysplasia involving the jaws alone. In 24 patients with severe polyostotic craniomaxillofacial fibrous dysplasia, early radical excision was performed with primary reconstruction. Mild forms of the disease affecting the maxilla, mandible, or both jaws in 19 patients were treated with conservative surgery and observed. Seven patients did not warrant any surgery and were placed under regular follow-up. In patients with fibrous dysplasia affecting Zone 4 alone, conservative excision and recontouring was performed. With early radical excision and reconstruction in polyostotic disease, 4 patients experienced dentofacial deformities severe enough to warrant orthognathic surgery. Orthognathic surgery was also indicated for only 1 patient with isolated jaw disease; the rest only required conservative excision and close observation.

Results

Between 1988 and 1997, orthognathic surgery was performed on 1 male and 4 female patients with fibrous dysplasia involving the teeth-bearing jaws (Zone 4) (Table). The patients ranged in age from 20 years to 37 years at the time of surgery. Only 1 patient had localized fibrous dysplasia that involved the mandible (Zone 4B). The other 4 patients had polyostotic craniofacial involvement of Zones 1, 4A, and 2 or 3. These 4 patients did not have any mandibular involvement. One patient with isolated mandibular involvement and 2 with maxillary fibrous dysplasia had single-jaw surgery. The other 2 patients with maxillary involvement required simultaneous two-jaw surgery to correct the dentofacial deformities resulting from the disease process. Follow-up ranged from 1 to 9 years. There was no mortality or morbidity associated with orthognathic surgery in these patients with craniomaxillofacial fibrous dysplasia. All the patients had stable occlusion, improved facial aesthetics, and no further recurrence after surgery (Fig 1). The long-term stability of the achieved occlusion and facial appearance confirmed adequate healing in fibrodysplastic bone using standard plate and screw fixation techniques.

Table. Orthognathic Surgery in Craniomaxillofacial Fibrous Dysplasia

Patient No.	*Age (years)/Sex*	*Diagnosis*	*Procedure*
1	13/M	FD of the (L) fronto-orbito-zygomatic, maxilla, hard palate and vomer Inferiorly displaced (L) maxilla, incisional overbite, interincisal diastema FD (L)	Subtotal excision FD; reconstruction with CBG; (L) hemi-Lefort 1 with 4 mm intrusion and 12 mm medial displacement, excision of the remaining FD
2	31/F	Isolated mandibular FD with asymmetrical prognathism, AXB, and rounded and protrusive chin	BSSO with 10 mm setback, and advancement genioplasty 6 mm; shaving inferior border
3	27/F	FD (L) maxilla, zygoma, and base of skull Incisional overbite of 5 mm and an overjet of 4.5 mm	Subtotal resection maxilla and zygoma and reconstruction with rib Lefort 1 with left maxillary wedge resection of 5 mm
4	35/F	FD (L) fronto-orbito-zygomatico-maxilla with optic nerve compression Class II occlusion, with 8.5 mm overbite and 11 mm overjet; midline shifted right 7 mm	Decompression and shaving ostectomy left zygoma and maxilla 3 piece Lefort 1 with intrusion (L) maxilla and resection FD; BSSO to level occlusal plane
5	31/F	FD (L) fronto-orbito-zygomatico-maxilla, optic nerve compression; 6 × 8 cm ulcer left NOE region; persistent maxillary excess and an anterior open bite	Excision FD with nerve decompression; reconstruction with bone grafts Lefort 1 (12 mm intrusion), BSSO (4 mm) setback and advancement genioplasty (8 mm)

FD = fibrous dysplasia; AXB = anterior crossbite; NOE = naso-orbitoethmoidal; CBG = cranial bone graft; BSSO = bilateral sagittal split osteotomy.

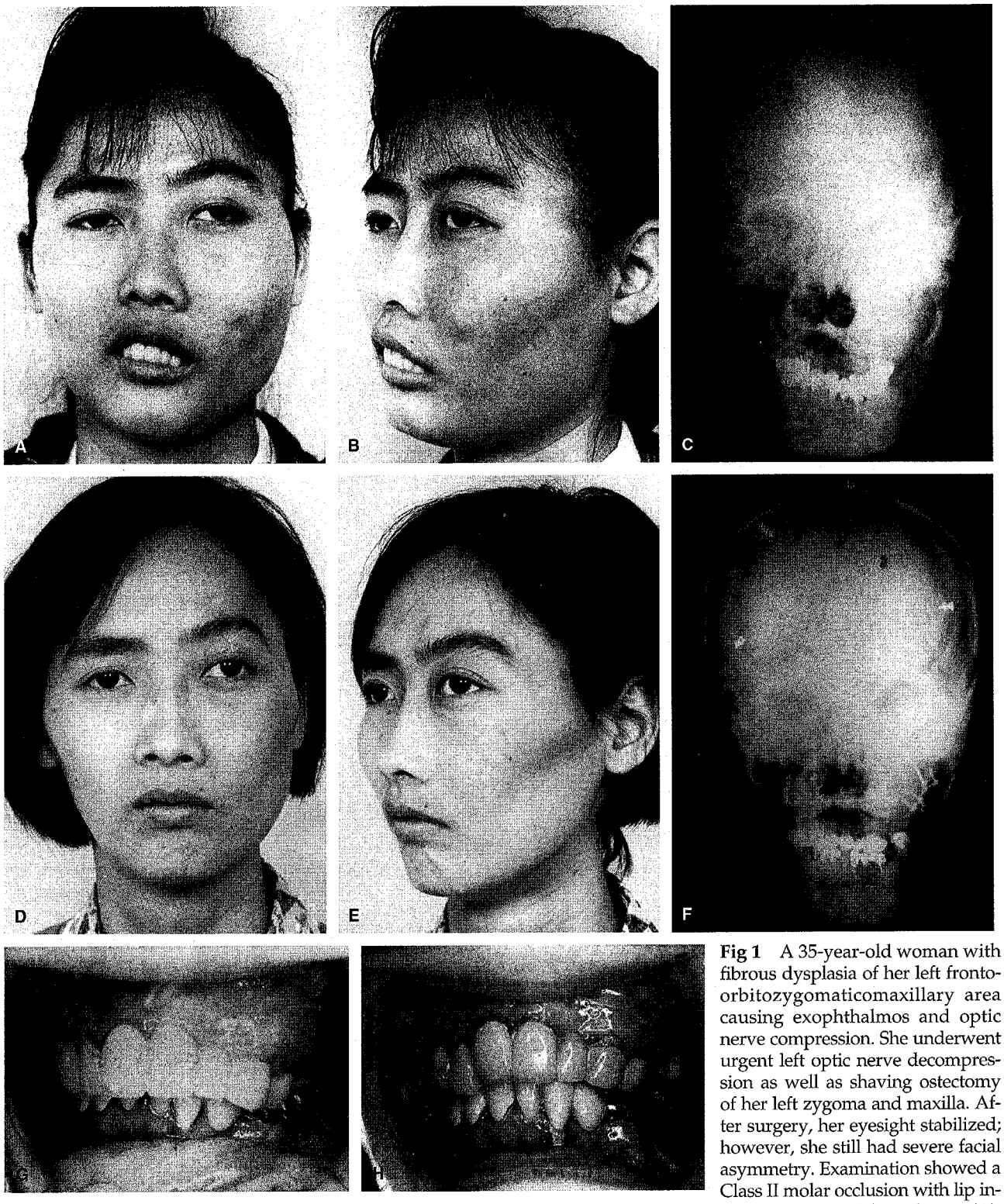

Fig 1 A 35-year-old woman with fibrous dysplasia of her left fronto-orbitozygomaticomaxillary area causing exophthalmos and optic nerve compression. She underwent urgent left optic nerve decompression as well as shaving ostectomy of her left zygoma and maxilla. After surgery, her eyesight stabilized; however, she still had severe facial asymmetry. Examination showed a Class II molar occlusion with lip incompetency. The maxillary midline was shifted to the right 7 mm with a resultant occlusal cant. Presurgical frontal view (A), angled view (B) and anteroposterior cephalogram (C). She underwent a 3-segment LeFort 1 osteotomy with intrusion of the left maxilla after radical wedge resection of the dysplastic bone and primary reconstruction with iliac bone grafts, a bilateral sagittal split of the mandibular rami to level the occlusal plane, and shaving of herleft orbitozygomatic fibrous dysplasia. Frontal view (D), angled view (E), and anteroposterior cephalogram (F) 3 years postoperatively. Presurgical occlusion (G) and stable postoperative occlusion 3 years postoperatively (H).

Discussion

Although fibrous dysplasia affecting the frontal, orbital, nasal, ethmoid, zygomatic, and upper maxillary region (Zone 1) of the face is now treated by early radical excision and primary reconstruction using bone grafts,[4] the teeth-bearing bones of the face are not amenable to this approach. Despite reservations about the unpredictability of recurrences[5] and the possibility of fibrous dysplasia occurring in reconstructed bone grafts,[6] the actual reason is that such radical surgery is essentially more destructive than in other parts of the facial skeleton because dentures will be required. Such dentures will never be as functional as natural teeth and, therefore, should be avoided unless malignant change dictates the need for radical surgery. Therefore, conservative surgery of shaving and recontouring is still the essence of surgical management in fibrous dysplasia of the jaws (Zone 4). In the course of the disease, dentofacial deformities may arise as a result of the progressive distortion of the mid and lower facial skeleton caused by the dysplastic bone. These deformities require orthognathic surgery to correct both dental and skeletal relationships.

In isolated single-jaw involvement, orthognathic surgery can be performed as the primary and principle procedure to correct the dentofacial deformity and malocclusion. This is usually combined with conservative excision and recontouring of the alveolus. Polyostotic craniomaxillofacial fibrous dysplasia, however, often results in gross craniofacial deformities of the lower, mid, and upper face. In such patients, the primary concern is often optic nerve decompression and radical excision of the fibrous dysplasia involving the frontal, orbital, nasal, ethmoid, zygomatic, and upper maxillary regions (Zone 1) with primary reconstruction using autogenous bone grafts. Once the upper and midface have been corrected, the dentofacial deformity can be dealt with at a second stage. The reconstructed upper and midface provide the necessary "normal" framework on which to base the planning and ultimately the osteotomies of orthognathic surgery to deal with the facial asymmetry and malocclusion. Surgical recontouring and conservative excision are again combined with the orthognathic procedure to maximize the aesthetic and functional results.

Sachs[7] and Samman[8] and their colleagues treated associated facial deformity caused by polyostotic fibrous dysplasia in a conservative manner. In their patients, they only performed shaving and recontouring of the affected zygoma and upper maxilla. From our experience, such a conservative approach is indicated only when either the disease process affects Zone 4 alone, causing mild asymmetry, or the fibrous dysplasia of the upper and midface is mild and quiescent. In our series, most of our patients with facial deformity affecting Zone 1 have polyostotic disease causing obvious facial deformity and often associated functional problems, such as deteriorating sight and obstructed nasal airways. The treatment of choice, therefore, is early primary radical excision and reconstruction. Despite reports of unpredictability of recurrences[5] and the possibility of fibrous dysplasia occurring in reconstructed bone grafts,[6] our previous experience[4] has shown stable reconstructed bone with only one incidence of recurrence in 24 patients. This patient had a large pituitary adenoma that was secreting excessively high levels of growth hormone. The patient's unusual clinical course was deemed to be caused by the pituitary adenoma.[9] Once the adenoma was excised, the fibrous dysplasia again became quiescent. In addition to the extirpation of the dysplastic bone, radical excision removes the deformational forces of the midface that will, if unchecked, cause subsequent dentofacial deformity and malocclusion.

Osteotomies performed in fibrodysplastic long bones have been reported by Freeman[10] and Grabias[11] and their associates with satisfactory healing. Orthognathic surgery for dentofacial deformities resulting from craniomaxillofacial fibrous dysplasia has been previously reported by Sachs and associates[7] in 1984, Samman and coworkers[8] in 1991, and Cheung and colleagues[12] in 1995. Our intraoperative findings are similar to those described by Cheung and others.[12] LeFort I osteotomies require sectioning through solid bone mass because the maxillary sinuses are obliterated by the fibro-osseous diseased bone. The extension of the dysplastic bone posteriorly to the skull base also requires one to be very careful with the downfracture as well as intrusion of the maxillary segment. Absolute care must be taken to preserve the vascular pedicles. Although Sachs and others[7] expressed uncertainty about segmentalizing the maxilla, standard segmentalization of the maxilla can be performed simultaneously, as demonstrated in our experience and in that reported by Cheung and colleagues.[12] Similarly, the mandible is thickened, and safe sagittal splitting is more difficult because the plane between the outer cortex and the cancellous bone is ill defined. Debulking and recontouring of the alveolus is accomplished by using an osteotome and shaving off slices of dysplastic fibro-osseous tissue down to bone just superficial to the tooth roots. Any irregularities can be smoothed with

a bur drill. This adds the finishing touch to the procedure and helps to maximize the aesthetic results.

Previous work by Cheung and coworkers[12] has already highlighted the "adequate biocompatibility of titanium screws with fibrous dysplasia in the long term." Fibrous dysplasia of the craniofacial region has been shown to be more osseous in nature when compared to FD in the long bones. Jackson and associates[13] in 1982 reported that craniofacial fibrous dysplasia was more fibro-osseous in nature, with bony trabeculae dominating. This has been verified histologically by Slootweg and Muller[14] as well as Waldron[15] in 1985. These researchers stated that, unlike the classical histological picture of fibrous dysplasia in long bones characterized by whorled connective tissue containing irregular woven bony trabeculae, the presence of lamellar bone and osteoblasts does not preclude the diagnosis provided the clinical, radiological, and operative picture was compatible with fibrous dysplasia. Therefore, the more ossified nature of the disease in the craniofacial area promotes both rigid fixation with plates and screws as well as the semirigid fixation afforded with wires combined with intermaxillary fixation. The long-term stability of the achieved occlusion and facial appearance in our series confirms that adequate healing in fibro-dysplastic bone is to be expected using the standard fixation techniques available to us today.

The management of dentofacial deformities caused by craniomaxillofacial fibrous dysplasia, therefore, requires orthognathic surgery and is dependent on the degree of deformity and functional disturbance. Early excision of midface disease with primary reconstruction provides the necessary "normal" architecture on which to base the planning of subsequent orthognathic surgery. In addition, radical excision removes the deformational forces of the midface that may cause subsequent dentofacial deformity and malocclusion. Conservative excision is often combined with the orthognathic procedure to maximize the aesthetic and functional results.

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